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TITLE: OTOLARYNGOLOGIC CLINICS OF NORTH AMERICA
PUBLISHER/PLACE: W B Saunders Philadelphia Pa
VOLUME/ISSUE/PAGES: 1988 Feb;21(1):103-18   103-18
DATE: 1988
AUTHOR OF ARTICLE: Stefanyszyn MA; Handler SD; Wright JE
TITLE OF ARTICLE: Pediatric orbital tumors.
ISSN: 0030-6665
OTHER NOS/LETTERS: Library reports holding volume or year
                      0144042
                      2829096
SOURCE: PubMed
CALL NUMBER: W1 OT518
REQUESTER INFO: AB424
DELIVERY: E-mail: probey@DIR.NIDCR.NIH.GOV
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Laryngoscope 83:945–959, 1973
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7:276–285, 1973
2:12–24, 1979
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Communication

Pediatric Orbital Tumors

Mary A. Stefanyszyn, MD, Steven D. Handler, MD,† and*
John E. Wright, MD‡

The appearance of an orbital mass in a child can be a frightening experience for parent and child alike. Proper management of these lesions requires a thorough knowledge of the types of orbital tumors seen in the pediatric age group (Table 1), the methods of detecting and evaluating local and distant disease, and the appropriate treatment. Some of these lesions may require a multidisciplinary approach, with the cooperation of the pediatrician, ophthalmologist, orbital or oculoplastic surgeon, otolaryngologist, plastic surgeon, neurosurgeon, and oncologist.

CONGENITAL DEVELOPMENT CYSTS

Dermoid and Epidermoid Cysts

Dermoid cysts present in the preschool child as painless, elevated nodules, most frequently seen along the superotemporal orbital rim (Fig. 1). However, they can occur superonasally or deep within the orbit. The posterior dermoids present at an older age, as they grow and produce proptosis. Dermoids are thought to occur as a result of entrapped epidermal tissue along bony sutures during embryonic development.[12, 14] The epidermal anlage develops into a cyst, lined by keratinizing epidermis with a stalk, usually to the zygomaticofrontal or nasofrontal suture. If the cyst wall contains adnexal structures, such as hair follicles, sweat glands, or sebaceous glands, it is referred to as a dermoid; if no adnexal structures are found in the cyst wall, it is considered an epidermoid. On CT scanning the cyst has a lucent low-density lumen, and in longstanding cases can display bony erosion secondary to cyst expansion (Fig. 2). Treatment consists of complete cyst removal, usually in the

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Table 1.   *Orbital Tumors in Children*

Congenital Development Cysts	*Fibro-osseous Tumors*
Dermoid	Fibrous dysplasia
Teratoma	Ossifying fibroma
Microphthalmos with cyst	Osteoma
Vascular Tumors	*Metastatic Tumors*
Capillary hemangioma	Neuroblastoma
Lymphangioma	Ewing's sarcoma
Mesenchymal Tumors	Leukemia
Rhabdomyosarcoma	
Neural Tumors	
Glioma	
Neurofibroma	

preschool years. Cyst rupture or incomplete removal can result in a lipogranulomatous inflammatory reaction, which is difficult to treat.[19]

Teratoma

Teratoma, a congenital tumor arising from aberrant germ cells, presents at birth with dramatic exophthalmos (Fig. 3).[1] Although benign, orbital teratomas are massive and can extend intracranially.[19] Histopathologically, all three embryonic layers—ectoderm, endoderm, and mesoderm—are represented. Teratomas are cystic; they contain numerous epidermoid cysts and embryonic forms of mucin-secreting gastrointestinal mucosa. CT scanning reveals a heterogeneous lesion with
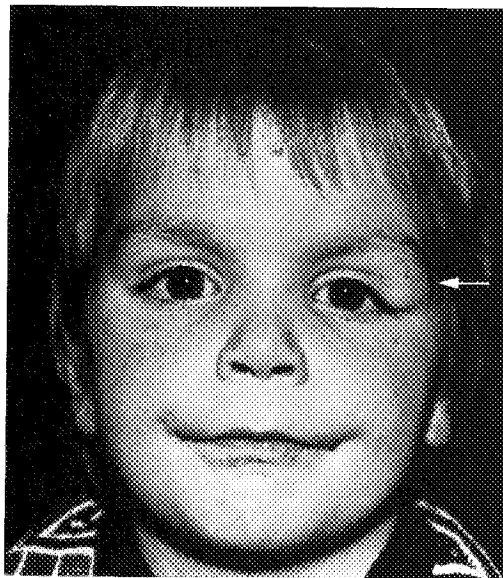
Figure 1.   Dermoid cyst located superotemporally along the left orbital rim, with a stalk to the zygomaticofrontal suture.

Fibro-osseous Tumors
Fibrous dysplasia
Ossifying fibroma
Osteoma

Metastatic Tumors
Neuroblastoma
Ewing's sarcoma
Leukemia

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c forms of mucin-secreting gas-
ls a heterogeneous lesion with

Figure 1. Dermoid cyst lo-
cated superotemporally along
the left orbital rim, with a stalk to
the zygomaticofrontal suture.

Figure 2. CT scan of a long-
standing dermoid cyst of the right
orbit. Note bony erosion and
fossa formation.
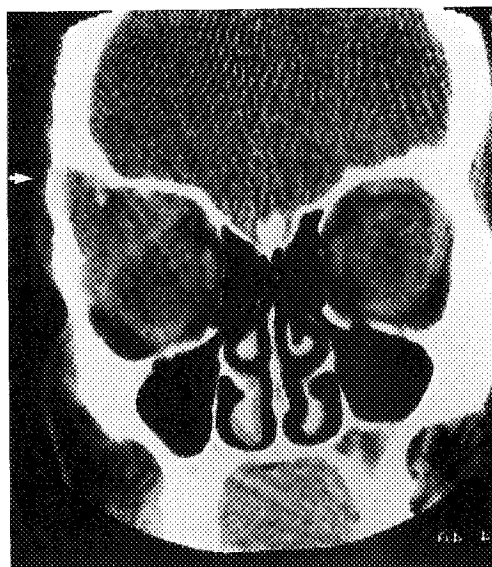
many cystic cavities and intracranial involvement (Fig. 4). Treatment
consists of a combined neurosurgical and orbital approach immediately
after birth, with preservation of the globe whenever possible.[3, 14, 17]

Microphthalmos with Cyst

A failure of closure of the embryonic fissure can result in a cystic
prolapse of the neuroretinal tissue into the orbit from a nonfunctioning
globe.[36] This condition is usually unilateral and the cyst can enlarge,
causing proptosis and sometimes obscuration of the microphthalmic

Figure 3. Orbital teratoma
presenting at birth with dramatic
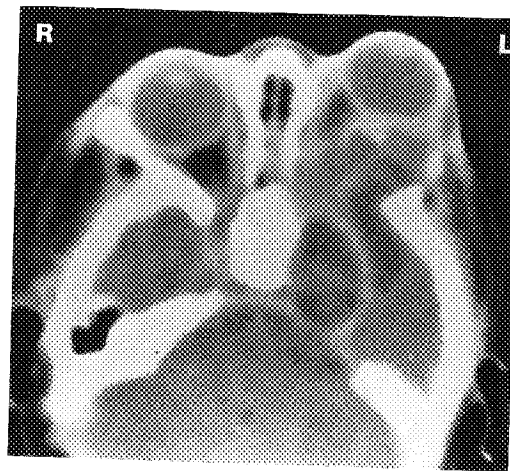exophthalmos (Courtesy of F.A.
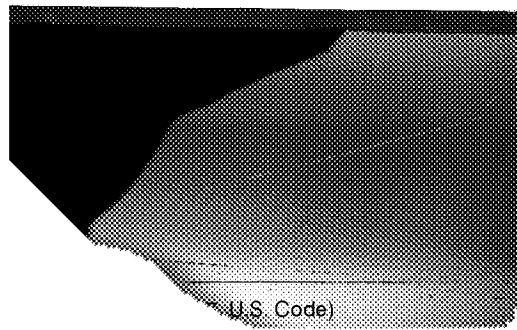Jakobiec, M.D., New York City.)

Figure 4. CT scan of orbital teratoma. Note the defect in the left sphenoid bone, with extension of the heterogeneous, retrobulbar, multicystic mass lesion into the temporal globe (Courtesy of D.H. Birstadt, M.D., Springfield, Illinois.)

globe. If the globe is totally microphthalmic, with no visual potential, an enucleation and excision of the cyst can be performed at the same time. The socket can then be reconstructed with a ball implant. If the globe is normal in appearance, or has some visual potential, an attempt should be made to excise the cyst, close the scleral defect, if any, and preserve the globe.

VASCULAR TUMORS

Capillary Hemangioma

Capillary hemangiomas develop within the first few weeks of life, and continue to grow until 2 or 3 years of age, at which time they spontaneously begin to involute. Although benign, capillary hemangiomas are invasive, unencapsulated, and hypercellular. They consist of lobular sheets of endothelial cells that surround small, capillary-like vascular spaces. The superficial lesions have the typical reddish strawberry appearance, whereas the deeper lesions can present with proptosis or eyelid swelling without the overlying skin changes. When the deeper lesions evolve rapidly, they can simulate malignancies such as rhabdomyosarcoma, and biopsy is indicated (Fig. 5). The upper eyelid is most commonly involved, resulting in amblyopia from occlusion of the pupil or from induced refractive error (Fig. 6).[8,31] In rapidly proliferating lesions, ulceration, bleeding, necrosis, and infection can occur. As these lesions involute, after their initial growth spurt, conservative management is indicated. However, if the lesion creates a severe cosmetic blemish, causes amblyopia, traps platelets causing a coagulopathy, causes high output cardiac failure, or is necrotic or infected secondary to breakdown of overlying skin, then treatment is indicated. Diffuse lesions are treated with systemic steroids or intralesional steroid injections.[2,13,26] In rare cases, excision of circumscribed lesions in the anterior orbit, or ligation or embolization of major arterial feeders, is indicated.[8,22]
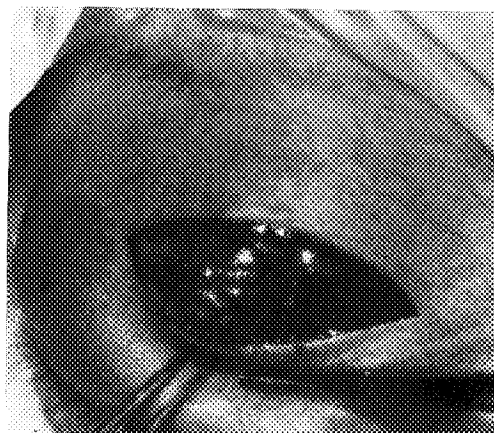
Figure 4. CT scan of orbital teratoma. Note the defect in the left sphenoid bone, with extension of the heterogeneous, retrobulbar, multicystic mass lesion into the temporal globe (Courtesy of D.H. Birstadt, M.D., Springfield, Illinois.)

Figure 5. Superficial biopsy of a capillary hemangioma to rule out malignancy.

Lymphangioma

Lymphangiomas are first noted in the preschool years. They present as soft, bluish masses, usually in the superonasal quadrant, and often have a cystic conjunctival component (Fig. 7). Posterior tumors can lie dormant for long periods of time, presenting suddenly, with painful proptosis secondary to spontaneous hemorrhage within the lesion. Histopathologic examination reveals attenuated channels lined by endothelial cells and filled with serous fluid or blood. Scattered lymphoid tissue is present in the stroma and may account for increased proptosis during infections. Debate continues as to whether this lesion is of venous or lymphatic

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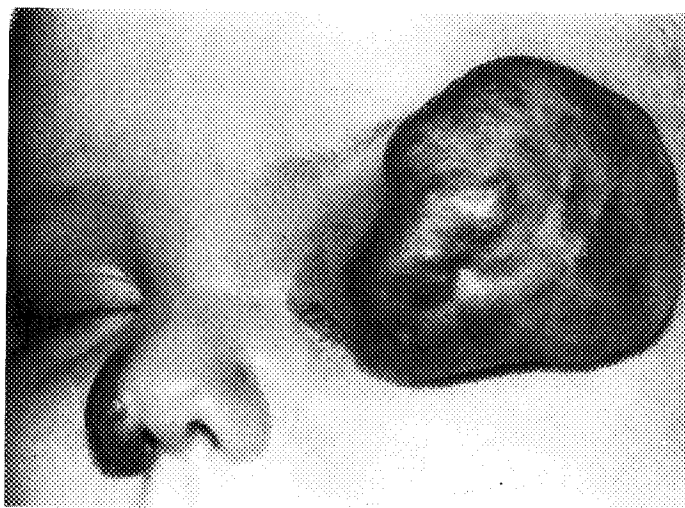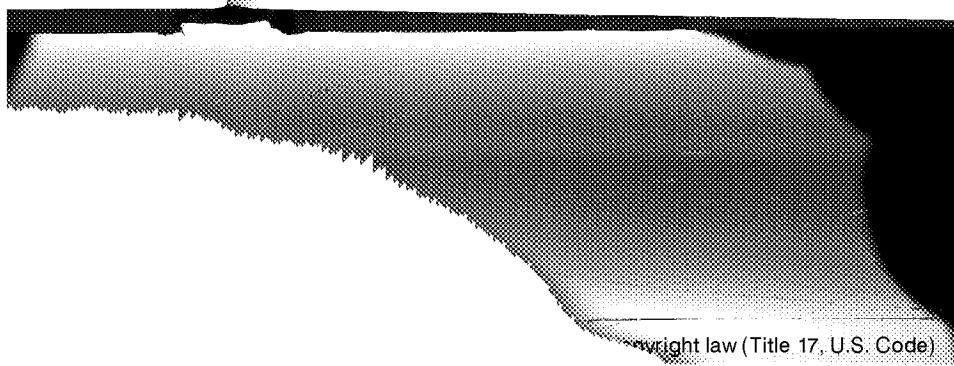feeders, is indicated.[8,22]

Figure 6. Typical strawberry-like capillary hemangioma of the left upper lid with occlusion of the visual axis. Note another capillary hemangioma on the nose.
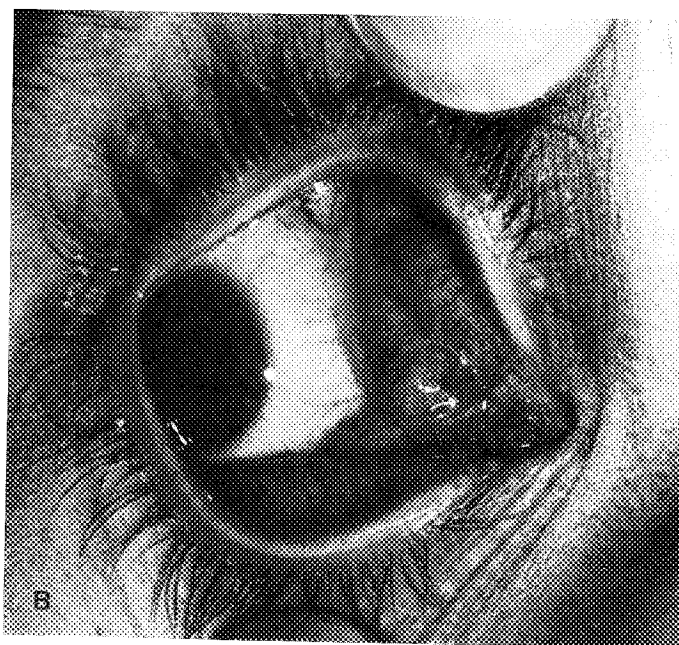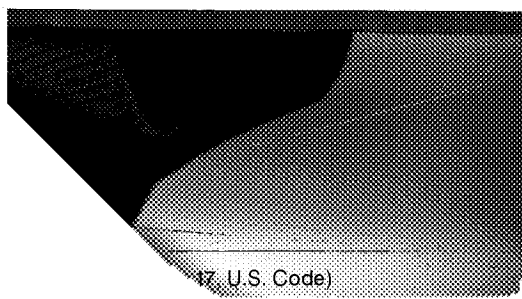
Figure 7. A and B, Lymphangioma involving the right superonasal orbit. Note the cystic conjunctival component.
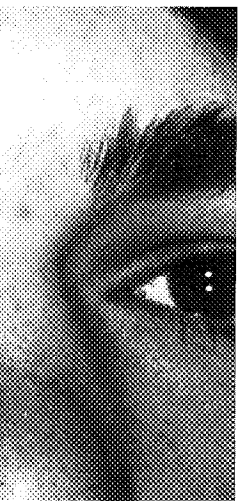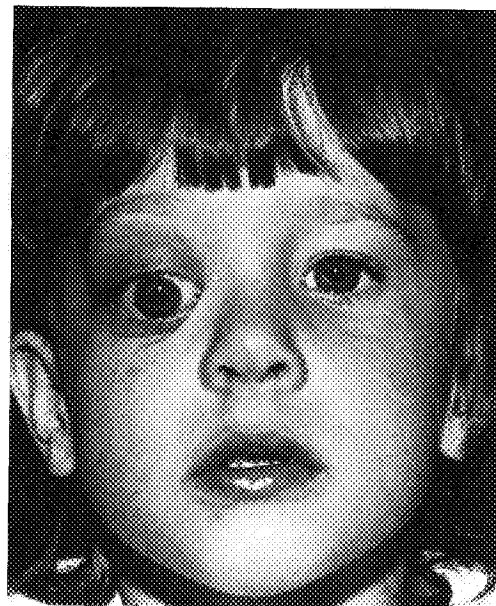
origin and perhaps is best referred to as vascular hamartoma.[18, 20, 39] Lymphangiomas are characterized by slow, indolent growth with occasional, acute exacerbations secondary to hemorrhage. This tumor is very difficult to excise surgically, because it is not encapsulated, infiltrates normal tissues, and has a tendency to bleed profusely.[21] Anterior debulking, with the help of a cutting cautery or carbon dioxide laser, or drainage of orbital hemorrhage, is indicated to correct severe cosmetic deformity and to prevent ocular dysfunction.[23] CT scanning is helpful in defining the posterior extent of these lesions and reveals their nonencapsulated, heterogeneous nature. Orbital lymphangiomas are often associated with venous dilations in the sinuses and oropharynx, which can result in nosebleeds.

MESENCHYMAL TUMORS

Rhabdomyosarcoma

Rhabdomyosarcoma is the most common childhood primary malignancy of the orbit. It usually presents as a rapidly evolving exophthalmos in children of approximately 7 years of age. The tumor most commonly involves the superior orbit (Fig. 8),[19] and can erode through the orbital bones into the sinuses, causing nasal stuffiness and nosebleeds. CT scanning reveals a poorly defined mass of homogeneous density, occasionally with areas of bone destruction (Figs. 9 and 10). An incisional biopsy, generally through an anterior approach, is performed with expediency. This is followed by a full systemic work-up, including physical examina-

he right superonasal orbit. Note the

Figure 8. Rhabdomyosarcoma of the right superior orbit, resulting in rapidly evolving proptosis, with downward displacement of the right globe.

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Figure 9. CT scan of rhabdomyosarcoma of the right orbit. Note the poorly defined mass filling the entire retrobulbar space.
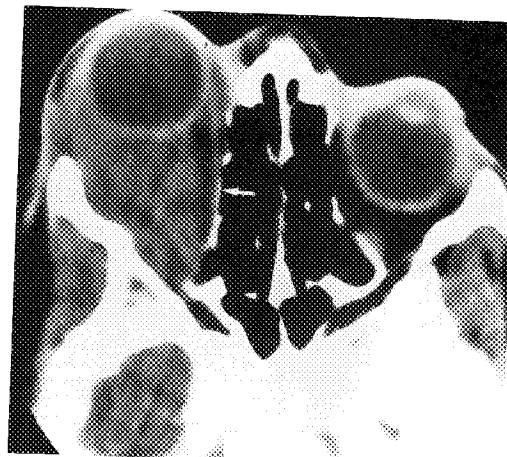
tion, chest radiograph, liver function tests, complete blood count, lumbar puncture, bone marrow biopsy, and skeletal radiographic survey.

Rhabdomyosarcoma can be of the embryonal, alveolar, botryoid, or pleomorphic type.[24] The embryonal form accounts for 80 per cent of pediatric cases and consists of loose fascicles of spindle cells, with eosinophilic cystoplasm. Trichrome stains can be helpful in demonstrating cross-striations of muscle filaments, and electron microscopy can demonstrate abortive sarcomeric organization with thick myosin and thin actin
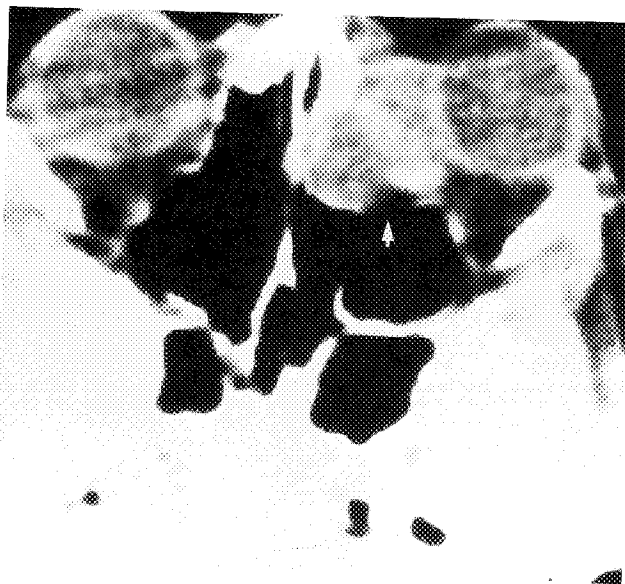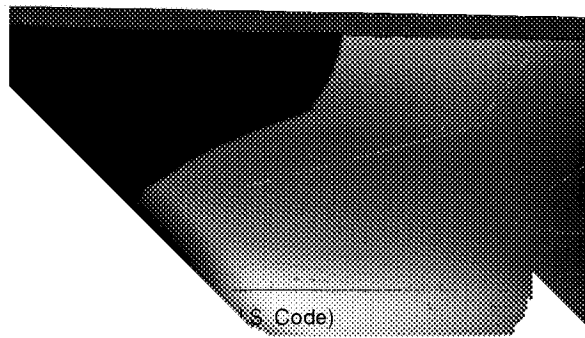

Figure 10. CT scan demonstrating destruction of the medial orbital wall and invasion of the ethmoid sinus by rhabdomyosarcoma.

filament. The alveolar form consists of rhabdomyoblasts arranged in an alveolar pattern. It is the second most common histologic form and has a predilection for the inferior orbit.[19] Once a definitive diagnosis is made, treatment consists of high-dose local radiation in the range of 5000 to 6000 rads. This is followed by systemic chemotherapy, including vincristine, adriamycin, and cyclophosphamide.[6] Treatment is best performed at an appropriate pediatric oncologic center. Exenteration is reserved for the rare radioresistant and recurrent tumor. Survival is now over 70 per cent. The most recent statistics from the Intergroup Rhabdomyosarcoma Study Committee reveal a 3-year survival of 93 per cent for children with localized orbital rhabdomyosarcoma.[37]

NEURAL TUMORS

Glioma

Gliomas, or juvenile pilocytic astrocytomas of the orbital optic nerve, present in preschool children with loss of vision, proptosis, papilledema, or optic atrophy and strabismus. The amount of proptosis can be minimal to massive, with an axial and downward displacement of the globe (Fig. 11). Twenty-five to 50 per cent of children with optic nerve gliomas have systemic neurofibromatosis.[27,35] CT scanning demonstrates a fusiform enlargement of the optic nerve, and optic canal views can show a concentric enlargement of the optic canal (Fig. 12). Magnetic resonance imaging (MRI) may be helpful in delineating the posterior extent of the tumor.

Treatment of orbital gliomas is controversial; some claim it is a benign hamartoma and should be treated conservatively. However, others believe it is an aggressive neoplasm and should be totally excised.[9,15,27,33] Wright and colleagues[40] have outlined the following schema for the management of optic nerve gliomas. If the tumor is re-

s, complete blood count, lum-
skeletal radiographic survey.
hbryonal, alveolar, botryoid, or
m accounts for 80 per cent of
les of spindle cells, with eosino-
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lectron microscopy can demon-
vith thick myosin and thin actin

n of the medial orbital wall and invasion

Figure 11. Axial and downward displacement of the left globe by an optic nerve glioma.
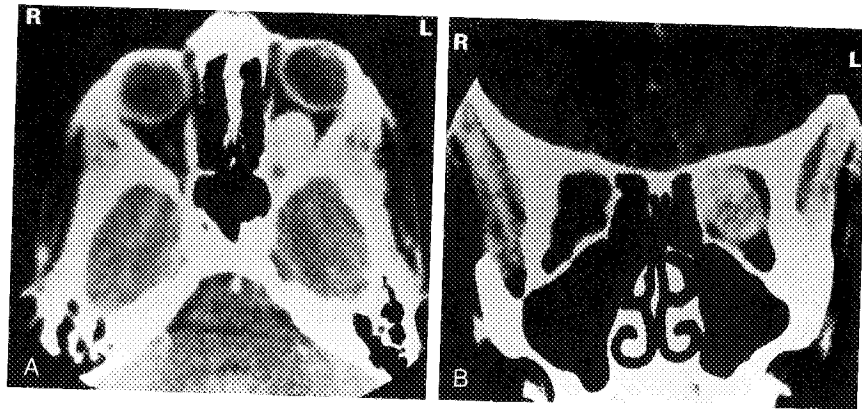
Figure 12. *A* and *B*, CT scan of an optic nerve glioma. Axial and coronal cuts demonstrate the fusiform enlargement of the optic nerve with a well-circumscribed homogeneous mass lesion.

stricted to the orbit, the vision is good, and there is minimal proptosis, then no treatment except careful observation, with periodic assessment of visual acuity, color vision, visual fields, and visual evoked response (VEP) is indicated. If vision is compromised, tumor growth is documented, and there is no involvement of the opposite optic nerve or chiasm, then surgical intervention should be contemplated. A combined neurosurgical and orbital approach allows visualization of the chiasm; and if it is not involved, the entire optic nerve, from the chiasm to the globe, can be removed. If chiasmal involvement is noted at the time of surgery, partial resection is performed for diagnosis and debulking.

Bilateral gliomas or those with chiasmal involvement are not treated surgically, with the exception of orbital resection for treatment of massive proptosis of blind, painful eyes or ventricular shunting for the relief of hydrocephalus. The efficacy of radiation therapy and chemotherapy for bilateral lesions is questionable.

Gliomas can grow within the optic nerve substance or they can grow circumferentially in a subdural location.[19, 34] Some assume a spindly, astrocytic pattern with eosinophilic inclusions called Rosenthal fibers; others are myxomatous with cystic areas that contribute to rapid tumor enlargement. Within the optic nerve, gliomas present with glial hypercellularity and disorderly arrangement. Reactive proliferation of adjacent arachnoidal cells can simulate a meningiomatous appearance and lead to errors on superficial biopsy.[4]

Neurofibromas

Neurofibromas can occur as solitary tumors or as a manifestation of systemic neurofibromatosis. In contrast to the solitary neurofibromas that occur past the second decade, the diffuse and plexiform lesions present in the early years. In the early stages, the plexiform neurofibromas present with ptosis and thickening, usually of the upper eyelid (Fig. 13). The tumor infiltrates all orbital tissues, making dissection difficult, if not impossible (Fig. 14).[25, 38, 41] The globe itself, including the

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Figure 13. Plexiform neu-
rofibroma presents with progres-
sive exophthalmos and ptosis of
the left upper lid.

sclera, iris, ciliary body, cornea, and choroid, can be infiltrated, resulting
in glaucoma and buphthalmos. The sphenoid bone may be partially de-
fective, resulting in pulsating exophthalmos. Treatment of this tumor
consists of multiple debulking procedures, which are often cosmetically
unsatisfactory. Visual loss occurs as a result of occlusion amblyopia, orbi-
tal and ocular infiltration, and glaucoma. The tumor is composed of
proliferating Schwann cells within their nerve sheaths, endoneural fibro-
blasts, and axons. Plexiform neurofibromas are diagnostic of neurofibro-
matosis. Other associated anomalies include cafe au lait spots, axillary
freckling, fibroma molluscum, dysplasia of the orbital walls, congenital
glaucoma, iris nodules, and optic nerve gliomas. The disease is inherited
through an autosomal dominant gene with irregular penetrance.

Figure 14. CT scan of a neu-
rofibroma with extensive in-
volvement of the orbital tissue.
Note dysplasia of the sphenoid
bone with intracranial extension
of tumor.
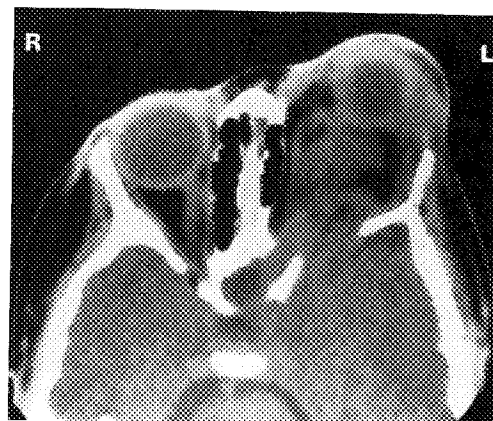
FIBRO-OSSEOUS TUMORS

There is a large group of loosely associated lesions of the orbit that are often grouped together under the heading of "fibro-osseous disorder." These include fibrous dysplasia, ossifying fibroma, lesions with giant cells (reparative granuloma, true giant-cell tumor, "brown" tumor of hyperparathyroidism) and osteoma.[10] The etiology of these lesions is obscure; trauma, infection, and developmental defects have all been considered as possible causes. All of these lesions are characterized by the replacement of normal bony architecture with tissue composed of varying amounts of collagen, fibroblasts, and osteoid and giant cells in differing patterns.

Fibrous Dysplasia

This is probably the most frequently seen lesion of this group and develops almost exclusively in children during the first 2 decades of life.[10,19,30] Two types of fibrous dysplasia have been described: polyostotic and monostotic. Polyostotic fibrous dysplasia (Albright's syndrome) consists of multiple bone involvement, abnormal skin pigmentation, and precocious puberty. The orbit is not generally involved in this systemic form of fibrous dysplasia. Monostotic fibrous dysplasia, on the other hand, occurs most often in the bones of the face and is the most common type seen in the orbit. Periorbital swelling or asymmetry are presenting signs of this lesion. The maxilla, sphenoid bone, or frontal bone are usually involved (Fig. 15). The radiographic appearance of this tumor is somewhat variable in that it depends upon the amount and distribution of osteoid matrix in the lesion. Generally, however, the radiographs in children with fibrous dysplasia show a sclerotic lesion with ground-glass appearance. Biopsy is usually necessary to confirm the diagnosis and to rule out a more aggressive lesion. On pathologic examination, the tissue shows bone trabeculae composed of woven bone within a fibrous stroma. This lesion is believed to represent an arrest in maturation of normal bone.[19] These lesions will often grow rapidly during early life and then stabilize after puberty. Conservative management is the hallmark of treatment. Small, stable lesions without any cosmetic or functional deformity require only observation.[10] Conservative surgical excision or sculpting has been the accepted treatment for lesions causing either cosmetic or functional (visual or sinonasal) symptoms. However, craniofacial surgeons have recently recommended radical excision of the diseased bone with immediate craniofacial reconstruction.[30] Radiation therapy does not appear to be efficacious, and has been implicated in the induction of malignant changes in fibrous dysplasia.

Ossifying Fibroma

This is often considered a variant of fibrous dysplasia. However, significant differences exist between these two lesions and warrant a separate category for each.[19,28,32] Ossifying fibroma is often characterized by a more aggressive growth pattern, often presenting with significant proptosis. The radiographic appearance of this lesion is quite distinctive in that it is more demarcated from normal bone than is fibrous dysplasia. Ossifying fibroma has a sclerotic margin surrounding a less

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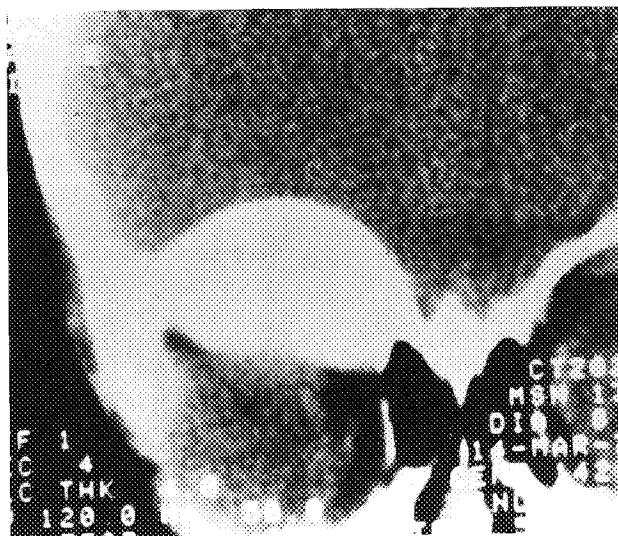tic margin surrounding a less

Figure 15. CT scan shows fibrous dysplasia of the right frontal bone resulting in sclerotic thickening of the orbital roof, resulting in proptosis and downward displacement of the globe.

radiodense center, which often contains foci of calcification. Because this is a more aggressive tumor, total surgical excision is the treatment of choice.[28, 32] The sharp demarcation that usually occurs between the ossifying fibroma and the surrounding normal bone makes total removal much more possible than in lesions of fibrous dysplasia. Pathologically, the ossifying fibroma consists of spicules of lamellar bone in a highly vascularized stroma. The spicules of bone are surrounded by osteoblasts and can be spheric in shape, thus resembling the psammoma bodies of meningioma.

Osteomas

These are uncommon, benign tumors of bone, occurring in the orbits of children. Radiologically, these lesions appear as well-circumscribed, extremely dense masses (Fig. 16). Simple surgical excision is the treatment of choice. Histopathologically, the osteoma is composed of compact bone without a fibrovascular stroma.

Giant-Cell Lesions

Lesions with giant cells include the giant-cell reparative granuloma, the true giant-cell tumor (rarely seen in children),[11] and the "brown" tumor of hyperparathyroidism. The radiographic appearance of these lesions is variable, depending upon the amount and configuration of matrix and osteoid. If a fibro-osseous lesion with giant cells is diagnosed, biochemical determinations (serum calcium, phosphorus, alkaline phosphatase, and parathormone) should be performed to rule out hyperparathyroidism as a cause.[10] Regression of the bony lesion often occurs after treatment of the hyperparathyroidism. Other lesions that contain giant cells usually respond to simple surgical curettage.[11]
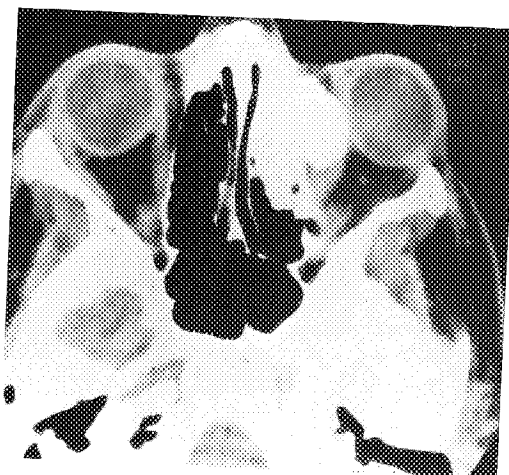
Figure 16. CT scan of an osteoma involving the ethmoid sinus, with extension into the medial orbital space.

METASTATIC ORBITAL TUMORS

The most common metastases to the orbit in the pediatric age group include neuroblastoma, Ewing's sarcoma, leukemia, medulloblastoma, and Wilms's tumor.

Neuroblastoma usually presents with ecchymotic unilateral or bilateral exophthalmos.[19] Metastases to the orbit occur with widespread disease in the abdomen, mediastinum, or neck. Metastases are usually to the lateral orbit, with bony destruction of the lateral wall. Ewing's sarcoma

Figure 17. Granulocytic sarcoma initially presenting with a mass in the superior orbit, without any evidence of systemic disease. Note downward displacement of the globe.

Figure 16. CT scan of an osteoma involving the ethmoid sinus, with extension into the medial orbital space.

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cchymotic unilateral or bilat-t occur with widespread dis-Metastases are usually to the ateral wall. Ewing's sarcoma

Figure 17. Granulocytic sarcoma initially presenting with a mass in the superior orbit, without any evidence of systemic disease. Note downward displacement of the globe.
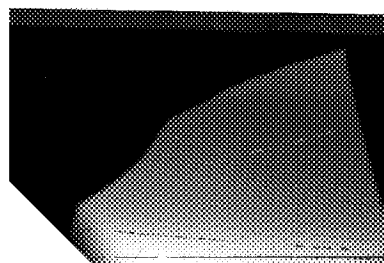
can present similarly with abrupt hemorrhagic exophthalmos, usually in the second decade. The primary lesions present in the long limb bones, truncal bones, metatarsal bones, or ribs.

Leukemic infiltrates to the orbit occur more frequently in the pediatric, rather than the adult, age group. Granulocytic sarcoma can present as a mass of infiltrating leukemic cells, without involvement of the peripheral blood and bone marrow (Fig. 17).[5] Clinically, this lesion has a greenish hue, and thus in the past has been referred to as a "chloroma." Histopathologically, this lesion is composed of immature cells, which stain positive for peroxidase granules by the Leder stain. Without treatment, overt leukemia develops within a period of 1 year. If chemotherapy is instituted prior to evidence of systemic disease, the prognosis is hopeful. When vision is acutely threatened or the orbital lesion is large, orbital radiotherapy can be efficacious.

CONCLUSION

The proper management of orbital tumors in children requires a knowledge of the lesions commonly seen in this area, in addition to the appropriate methods of investigation and treatment. A multidisciplinary approach, involving the otolaryngologist, ophthalmologist, radiologist, pathologist, and oncologist, is most beneficial in obtaining the desired results.

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